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Angioedema associated with monoclonal gammopathy

2/21/2011

Q. **Over the past few years, I have seen several mature patients (>70 yrs with angioedema without urticaria who are found to have a monoclo gammopathy. Usually these are determined to be MGUS, although or MM. The evaluation otherwise is negative with no complement abnor ACE inhibitors, NSAIDs, foods, etc. The patients usually respond pro steroids, etc. during episodes, but preventative therapy with antihist has been problematic. Is it possible that the monoclonal protein is a contact system to trigger attacks? If so, how is this evaluated in term studies, etc., and are these best performed during angioedema attac suggestions appreciated. Thank you.**

A. Thank you for your recent inquiry.

I have not had any experience with the type of patient you have described perhaps because I don't normally obtain an immunofixation, immunoelectr serum protein electrophoresis in patients over 70 years of age with angioe indeed may be a syndrome that I simply have not recognized. Of course, i due to the simple coincidence that MGUS occurs not infrequently at this a angioedema.

Nonetheless, since I cannot give you a definite answer, I am going to ask Dr. Allen Kaplan, who as you know is an internationally known expert in th angioedema. When I receive Dr. Kaplan's response, I will forward it to you

Thank you again for your inquiry.

Sincerely,
Phil Lieberman, M.D.

We received a response from Dr. Allen Kaplan, which is copied below. Th for your inquiry, and we hope this response is helpful to you.

Sincerely,
Phil Lieberman, M.D.

Response from Dr. Allen Kaplan:
When one sees angioedema in an older individual, in the absence of urtic C1 inhibitor deficiency comes to mind. Usually one would draw blood for a

would be quite low if that turns out to be the diagnosis, and the C1 Inhibitc
be abnormally low both by function and protein. Thus it looks like Heredita
biochemically. A C1Q level is low 70% of the time in the acquired disorder
distinguishes it from the hereditary disorder where C1Q is normal. When t
made, an underlying B-cell lymphoma may be present, but also possible i
gammopathy. If present, the abnormal protein is usually an antibody to the
The history you sent mentions absence of any complement abnormality w
what was drawn, so check to be sure the aforementioned tests were done
really no complement abnormality, them the diagnosis may be idiopathic a
someone who coincidentally has a monoclonal gammopathy. The only oth
of a monoclonal spike and related disorders is Schnitzlers Syndrome whic
urticaria/urticarial vasculitis rather than angioedema and lots of constitutio

Sincerely,
Allen Kaplan, M.D.

Key Words: angioedema, monoclonal gammopathy

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